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Title

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Permalink

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Journal

Pediatric neurology, 2(6)

ISSN

0887-8994

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Publication Date

1986-11-01

DOI

10.1016/0887-8994(86)90083-4

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Peer reviewed

Atypical Spasmus Nutans as an Initial Sign of Thalamic Neoplasm

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A patient is described who presented with dissociated nystagmus (atypical spasmus nutans) and an underlying pulvinar-tectal lesion. Atypical spasmus nutans is discussed and clinicians are alerted to a spectrum of possible etiologies.

Baram TZ, Tang R. Atypical spasmus nutans as an initial sign of thalamic neoplasm. *Pediatr Neurol* 1986;2:375-6.

Introduction

Spasmus nutans, a term first used by Raudnitz [1], was described initially in 1888 by Norrie [2]. It consists of head tilt, head nodding, and dissociated nystagmus. Onset usually occurs during the patient's first 18 months of life and the condition generally disappears by 3 years of age [3], although longer durations have been reported [4]. The nystagmus is usually fine, horizontal, pendular, rapid, and classically asymmetric or unilateral [5]. The head tilt and head nodding are variable and are not related to the nystagmus. Unlike congenital nystagmus, spasmus nutans head movements do not compensate for the nystagmus and may even suppress it [6]. Commonly, the first sign of the condition is the child's head tilt or head nodding. In classic spasmus nutans, the remainder of the patient's neurologic examination is unremarkable; with the exception of strabismus [4], there should be no evidence of visual abnormalities, field defects, or diencephalic syndrome. Thus defined, classic spasmus nutans rarely has been associated with underlying pathology [3,4].

Atypical spasmus nutans is used by several investigators to categorize dissociated nystagmus acquired in early childhood. There may be associated head tilt or head nodding, but the sequence of appearance or the



Figure 1. Contrast-enhanced computed tomogram depicting marginally enhancing thalamic mass. (Septum cavum pellucidum also is present).

age at presentation may be atypical. Several authors have reported the association of atypical spasmus nutans with lesions of the anterior optic pathway [7-9] or with the anterior third ventricular region [7,9].

Our patient had atypical spasmus nutans associated with an underlying primary thalamic lesion, a location not previously linked to this syndrome. We discuss the



Figure 2. T₁-weighted magnetic resonance imaging study, 5 mm slice, (TR = 1,000 msec, TE = 28 msec) revealing distortion of the quadrigeminal plate by a thalamic mass.

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Received August 28, 1986; accepted October 1, 1986.

validity of the term, "atypical spasmus nutans," and evaluate the possibility of an association with lesions of the posterior third ventricle.

Case Report

The patient, referred at age 10½ months, was the product of a 37 week gestation in a 28-year-old woman whose pregnancy was complicated by third-trimester pre-eclampsia. The delivery and perinatal period were uncomplicated. Developmental milestones were normal. There was no family history of nystagmus or brain tumors. Her unusual eye movements, observed initially at 6 months of age, consisted of dissociated horizontal nystagmus confined to the right eye. Referral to an ophthalmologist resulted in the diagnosis of a typical spasmus nutans. No head nodding or head tilt was present.

Examination showed the infant to be physically and developmentally normal. Her head circumference was at the fiftieth percentile. The abnormalities found during neurologic examination were limited to the extraocular movements; a dissociated horizontal mixed pendular-jerk nystagmus, confined to the right eye, was evident in all directions of gaze but maximal on right lateral gaze. On upgaze there was a rare, bilateral upbeat jerk nystagmus but no evidence of upgaze limitation or of any component of the dorsal midbrain (Parinaud) syndrome [10]. Cranial computed tomography revealed the presence of a slightly hyperdense, posterior thalamic (pulvinar) lesion with minimal enhancement on post-contrast study (Fig 1). The lesion extended to the midbrain tectum and to the roof of the third ventricle. Magnetic resonance imaging in the sagittal plane demonstrated the abnormally thickened tectum and deformed quadrigeminal plate (Fig 2).

Discussion

Spasmus nutans is a well-defined clinical entity, consisting of head nodding, head tilt ("torticollis"), and dissociated nystagmus which appear in the first or second year of life [4,5]. Classically, the condition is transient and not associated with any discernible underlying pathology [4]. Atypical spasmus nutans is often diagnosed when some of the features of the classic syndrome are absent or when the patient's age at presentation is inappropriate [9]. The ophthalmologic literature recognizes dissociated nystagmus alone or in combination with head tilt as atypical spasmus nutans [4,7]. In the absence of the classic triad of symptoms, atypical spasmus nutans has been reported frequently in association with anatomic lesions, specifically tumors involving the optic chiasm and hypothalamus [7-9,11]. Some authorities suggest applying the term pseudo-spasmus nutans to the entity of atypical spasms nutans

with a well-defined underlying pathologic process [D. G. Cogan, personal communication, 1986].

The precise mechanism by which a lesion in the area of the optic chiasm causes dissociated nystagmus with or without head tilt or nodding is unclear. Dissociated nystagmus can be induced by electrical stimulation of the superior colliculus [11]; clinically it is most frequently found with lesions of the medial longitudinal fasciculus (MLF) [5]. An MLF lesion also commonly underlies purely unilateral jerk nystagmus, contralateral to the lesion, with ipsilateral adduction palsy [5,10]. Stimulation of the quadrigeminal plate also can produce monocular nystagmus [12].

The lesion in our patient appeared to emanate from the posterior thalamus and impinge upon and deform the quadrigeminal plate, thereby resulting in the monocular nystagmus.

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